CASE REPORT

OSTEOPLASTIC MAXILLOTOMY APPROACH FOR INFRAORBITAL NERVE SCHWANNOMA, A CASE REPORT

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Abstract: Background. Extracranial schwannomas can readily occur in the head and neck region and rarely involve the trigeminal nerve. As a rule, their treatment is surgical and dictated by the location of the tumor and nerve of origin.

Methods. We describe a case of a 14-year-old boy with a mass invading right nasal fossa, maxillary sinus, orbital floor, pterygopalatine fossa, and infratemporal fossa. The diagnosis of a nerve sheath tumor was evoked after angiography showed no vascular blush.

Results. The tumor was removed through a Weber-Fergusson incision with subciliary extension followed by maxillozygomatic osteotomy. This approach showed the tumor to be coming from the infraorbital nerve and allowed complete tumor exposure and removal. Pathology confirmed the diagnosis of a schwannoma.

Conclusion. We describe the osteoplastic maxillotomy approach which we felt most appropriate for removal of the infraorbital schwannoma and discuss other possible surgical options for this type of tumor.

Keywords: head and neck schwannoma; infraorbital nerve; maxillary sinus; pterygopalatine fossa; infratemporal fossa

The treatment of extracranial head and neck schwannomas is surgical and the approach depends on the location and extent of the tumor and the nerve involved.

We report a rare case of a schwannoma involving the infraorbital branch of the trigeminal nerve in a 14-year-old male adolescent. We describe our surgical technique for tumor resection and discuss other possible surgical approaches for this particular localization.

CASE REPORT

A 14-year-old boy was seen with headache and right nasal obstruction ongoing for 1.5 months; there was no associated rhinorrhea or epistaxis. Physical examination showed right exophthalmus and scleral show with bulging of the right cheek (Figure 1A). Anterior rhinoscopy and fiberoptic nasopharyngoscopy revealed a moderately vascularized mass obstructing the posterior half of the right nasal cavity. Examination of the oral cavity and oropharynx showed bulging of the hard palate on the right side. There was no hypoesthesia of the right hemiface.
Noncontrast CT scan showed a 59.5- × 43-mm isointense opacity occupying the posterior two thirds of the right maxillary sinus, with extension into the orbit through the orbital floor, the pterygopalatine fossa through the posterior wall of the maxillary sinus, the infratemporal fossa through the pterygoid plates and pterygomaxillary fissure, and the posterior half of the nasal cavity through the medial wall of the maxillary sinus. There was no bone erosion of the hard palate or the base of skull (Figure 2).

MRI showed the mass to be hypointense on T1-weighed signal and isointense on T2, with significant enhancement upon gadolinium injection. The orbital contents were pushed anteriorly and the soft tissues of the cheek slightly anterolaterally (Figure 3).

The initial impression was that the tumor may be a juvenile angiofibroma. However, angiography did not demonstrate any vascular blush, and therefore, a diagnosis of a nerve sheath tumor was suspected. Surgical resection through an osteoplastic maxillotomy approach was then planned.

Access to the tumor was obtained via a Weber-Fergusson incision (lateral rhinotomy with superior lip split) extended by a subciliary incision with lateral flap elevation followed by an en bloc osteoplastic maxillotomy and zygomatic osteotomy (Figures 4A and 4B).

This approach largely exposed the anterior wall of the tumor. Frozen section revealed a schwannoma. The infraorbital nerve was closely related to the tumor and probably the point of origin; it was thus sacrificed (Figure 4C). The tumor was firm and encapsulated and was occupying the posterior half of the right nasal fossa and maxillary sinus, the pterygopalatine fossa, the infratemporal fossa, and the lower part of the orbital cavity. It was easily removable except superiorly (orbit) and posteriorly (pterygopalatine fossa); at these levels, the tumor was removed using the microscope with very cautious dissection to avoid retaining any tumor residue. The destroyed orbital floor was reconstructed by a synthetic, nonresorbable plate, and the maxillozygomatic osteoplastic flap was placed back and held in place using plates and screws (Figure 4D).

Postoperatively, there were no visual disturbances except for a slight diplopia which disappeared completely 4 months after surgery. There was, however, a discrete “sinking” of the ipsilateral eye, ie, a depression at the level of the inferior orbital rim due to scarring of the skin overlying

**FIGURE 1.** (A) Patient at presentation; note right exophthalmus (arrowhead) and cheek swelling (arrow). (B) Patient approximately 4 months after surgery. Note discrete sinking of the right eye (arrow). Surgical scar is barely visible.

**FIGURE 2.** CT scan with a coronal view showing an opacity invading maxillary sinus (black star), floor of orbit (white star), and infratemporal fossa (arrow).

**FIGURE 3.** MRI image after gadolinium administration with a coronal view demonstrating marked enhancement of the tumor (black star).
the plate and loss of the inferior palpebral fat pad, probably secondary to ischemia of the adipose tissue; otherwise, the surgical scar was very discrete and barely visible (Figure 1B). On the other hand, anesthesia of the skin innervated by infraorbital nerve persisted until that date. Finally, a control MRI at 7 months from surgery showed no tumor residue (Figures 5A and 5B).

DISCUSSION

Schwannomas arise from Schwann cells present in nerve sheaths and grow very slowly in close relationship with the nerve of origin.1,2 Of the 12 cranial nerves, only the olfactory and optic nerves do not develop schwannomas since they lack Schwann cells in their sheaths.3,4

Between 25% and 45% of extracranial schwannomas occur in the head and neck region.3,4 Extracranial fifth cranial nerve involvement appears to be less frequent than cranial nerves IX, X, XI, and XII.2,4

CT scan clearly demonstrates the location and extent of tumor growth and bone changes.5 Schwannomas appear isodense or slightly hyperdense, uniformly enhancing after contrast injection.5,6 MRI is complementary to CT and offers major advantages including increased sensitivity in detecting contrast differences, displacement of normal structures, and vascular structures in relation to the lesion. There is a variable signal intensity on T1-weighed images and an increased signal intensity on T2 with significant enhancement after gadolinium injection.5,6 Angiography helps differentiate schwannomas from other more vascularized tumors (juvenile angiofibromas, glomus tumors) due to the absence of vascular blush.5,7

The treatment of extracranial head and neck schwannomas is exclusively surgical. In schwannomas arising from the infraorbital nerve sheath, the appropriate approach is dictated by the extension of the tumor. In our case, the tumor originated from the infraorbital nerve and invaded the maxillary sinus, pterygopalatine fossa, infratemporal fossa, and lower orbit; its extension was thus anteroposterior then posterolateral. The transfacial approach seemed appropriate for such a localization,5 and the tumor was widely exposed and completely removed. Another alternative would have been the limited transzygomatic or
limited rhytidectomy approach, which consists of a preauricular skin incision extended to the scalp, temporal flap elevation, division of the zygomatic arch and coronoid process with superior elevation of the temporalis muscle flap. This approach is usually indicated for upper infratemporal fossa tumors with medial extension into the orbit. Although the limited rhytidectomy incision is more discrete than the lateral rhinotomy, we thought that the latter was more appropriate since the infratemporal fossa was invaded by lateral extension of the tumor and was not the site of tumor origin and anterior exposure of the tumor would have been troublesome with the lateral rhytidectomy approach. Other larger approaches to the infratemporal fossa (extended rhytidectomy, infratemporal extension of lateral transparotid approach, lateral transmandibular approach) are too excessive and not indicated for an infraorbital nerve tumor extending into the infratemporal fossa. Access to the orbit is also limited via these lateral approaches. Conversely, more conservative median approaches (Caldwell-Luc, Rouge-Denker) are very limited and do not give adequate superior and lateral tumor exposure. They should be used only in case of very limited and anteriorly localized tumors: Katz et al (1971) successfully removed a 1.5-cm schwannoma of the infraorbital nerve not invading bone through a sublabial incision. Finally, 2 sublabial approaches could have been possible alternatives to the Weber-Fergusson incision with subciliary extension with the advantage of leaving no visible facial scar; nevertheless, they present significant disadvantages and did not seem adequate for our case. The first is bilateral sublabial incision followed by Lefort I maxillary osteotomy as described by Negrier et al (1985). Despite good exposure, this type of surgery is not devoid of postoperative morbidity and requires maxillary bone suspension and intermaxillary blockage for 3 weeks. The second is midfacial degloving consisting of a bilateral sublabial incision connected to intranasal circumvestibular incisions with midface flap elevation followed by medial maxillectomy. It offers good exposure to the nasal fossa, maxillary sinus, medial part of the orbit, and pterygopalatine fossa (by removing the posterior wall of the maxillary sinus). However, access to the lateral part of the orbit and the infratemporal fossa is very limited and would have not allowed complete tumor removal in our case.

CONCLUSION
We report on a rare case of infraorbital nerve schwannoma and describe the osteoplastic maxillotomy approach for removal of this tumor. Despite the presence of many other alternative techniques, our surgical approach seemed most appropriate for such a tumor localization; it provided very good exposure and complete resection of the tumor with minimal aesthetic compromise.

REFERENCES